Craniovertebral Junction Realignment for the Treatment of Basilar Invagination With Syringomyelia: Preliminary Report of 12 Cases

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Abstract

Twelve selected patients, eight males and four females aged 14 to 50 years, with syringomyelia associated with congenital craniovertebral bony anomalies including basilar invagination and fixed atlantoaxial dislocation, and associated Chiari I malformation in eight, were treated by atlantoaxial joint manipulation and restoration of the craniovertebral region alignment between October 2002 and March 2004. Three patients had a history of trauma prior to the onset of symptoms. Spastic quadriparesis and ataxia were the most prominent symptoms. The mean duration of symptoms was 11 months. The atlantoaxial dislocation and basilar invagination were reduced by manual distraction of the facets of the atlas and axis, stabilization by placement of bone graft and metal spacers within the joint, and direct atlantoaxial fixation using an inter-articular plate and screw method technique. Following surgery all patients showed symptomatic improvement and restoration of craniovertebral alignment during follow up from 3 to 20 months (mean 7 months). Radiological improvement of the syrinx could not be evaluated as stainless steel metal plates, screws, and spacers were used for fixation. Manipulation of the atlantoaxial joints and restoring the anatomical craniovertebral alignments in selected cases of syringomyelia leads to remarkable and sustained clinical recovery, and is probably the optimum surgical treatment.

Key words: atlantoaxial dislocation, basilar invagination, Chiari malformation, syringomyelia

Introduction

The complex of basilar invagination, Chiari I malformation, and syringomyelia is relatively common, whereas the association with fixed atlantoaxial dislocation is less common but not rare.\(^1\),\(^4\),\(^7\),\(^13\),\(^15\)–\(^20\)

Such cases are generally treated by either anterior transoral or posterior foramen magnum bony decompression. The indications for opening of the dura and manipulation of the arachnoid membrane, tonsils, and obex, and draining of the syrinx cavity are currently under discussion. No specific fixation procedure has been recommended for fixed atlantoaxial dislocation associated with basilar invagination in the presence of syringomyelia.\(^9\)

We previously classified cases of syringomyelia into three groups and suggested specific treatment protocols on the basis of the possible pathogenetic factors.\(^7\),\(^8\) We suggested that syringomyelia is a tertiary response to reduction in the posterior cranial fossa volume resulting from the primary craniovertebral anomaly of basilar invagination and secondary Chiari I malformation. Accordingly, posterior fossa bony decompression was recommended for this subgroup of patients.

The present study included 12 patients with syringomyelia associated with bony abnormalities of the craniovertebral region including ‘fixed’ atlantoaxial dislocation and basilar invagination, and with Chiari I malformation in eight patients. All patients were treated by attempted reduction of the atlantoaxial dislocation and basilar invagination, and by direct lateral mass plate and screw atlantoaxial fixation by our reported techniques.\(^5\),\(^6\),\(^11\) No bony or dural decompression or neural manipulation of any kind was performed.

Patients and Methods

Twelve patients with syringomyelia associated with ‘fixed’ atlantoaxial dislocation and basilar invagina-
Table 1  Principal presenting clinical features

<table>
<thead>
<tr>
<th>Clinical features</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Preoperative</td>
</tr>
<tr>
<td>Neck pain</td>
<td>9</td>
</tr>
<tr>
<td>Torticollis</td>
<td>3</td>
</tr>
<tr>
<td>Lower cranial nerve affection</td>
<td>1</td>
</tr>
<tr>
<td>Weakness</td>
<td></td>
</tr>
<tr>
<td>able to walk unaided</td>
<td>10</td>
</tr>
<tr>
<td>needed support to walk</td>
<td>2</td>
</tr>
<tr>
<td>Sensations</td>
<td></td>
</tr>
<tr>
<td>normal sensations</td>
<td>3</td>
</tr>
<tr>
<td>kinesthetic sensations affected</td>
<td>6</td>
</tr>
<tr>
<td>spinothalamic sensations affected</td>
<td>9</td>
</tr>
<tr>
<td>Sphincter disturbance</td>
<td>2</td>
</tr>
</tbody>
</table>

Table 2  Preoperative radiological measurements and changes following surgery

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age/Sex</th>
<th>CL (mm)</th>
<th>WL (mm)</th>
<th>ADI (mm)</th>
<th>Omega angle (degrees)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Pre</td>
<td>Post</td>
<td>Dt</td>
<td>Pre</td>
</tr>
<tr>
<td>1</td>
<td>22/M</td>
<td>15</td>
<td>12</td>
<td>3</td>
<td>6</td>
</tr>
<tr>
<td>2</td>
<td>23/M</td>
<td>8</td>
<td>4</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>3</td>
<td>47/M</td>
<td>8</td>
<td>4</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>4</td>
<td>48/M</td>
<td>13</td>
<td>4</td>
<td>9</td>
<td>8</td>
</tr>
<tr>
<td>5</td>
<td>14/M</td>
<td>9</td>
<td>6</td>
<td>3</td>
<td>15</td>
</tr>
<tr>
<td>6</td>
<td>30/M</td>
<td>8</td>
<td>5</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>7</td>
<td>37/M</td>
<td>10</td>
<td>4</td>
<td>6</td>
<td>8</td>
</tr>
<tr>
<td>8</td>
<td>50/F</td>
<td>14</td>
<td>8</td>
<td>6</td>
<td>11</td>
</tr>
<tr>
<td>9</td>
<td>35/F</td>
<td>8</td>
<td>3</td>
<td>5</td>
<td>9</td>
</tr>
<tr>
<td>10</td>
<td>24/F</td>
<td>12</td>
<td>7</td>
<td>5</td>
<td>10</td>
</tr>
<tr>
<td>11</td>
<td>26/M</td>
<td>18</td>
<td>7</td>
<td>11</td>
<td>7</td>
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<tr>
<td>12</td>
<td>32/F</td>
<td>14</td>
<td>8</td>
<td>6</td>
<td>12</td>
</tr>
</tbody>
</table>

Values show the distance between the tip of the odontoid process and the anatomical references. Chamberlain’s line (CL): This line connects the posterior border of the hard palate to the opisthion. The tip of the odontoid process normally lies below this line. Wackenheim’s line (WL): This line is the clivus baseline extended downwards. Normally, the tip of the odontoid process remains anterior to this line. Modified omega angle: A line is drawn traversing along the center of base of the axis parallel to the line of the hard palate. The inclination of the tip of the odontoid process to this line is defined as the modified omega angle. ADI: atlantodental interval, Alt: alteration, Dt: distraction, Pre: preoperative, Post: postoperative.

Craniovertebral Realignment for Syringomyelia were treated between October 2002 and March 2004. Eight patients also had Chiari I malformation. Atlantoaxial dislocation was defined as ‘fixed’ if the minimum distance between the tip of the odontoid process and the posterior surface of the anterior arch of the atlas or clivus (in patients with assimilated atlas) was more than 4 mm and there was no clear radiographic evidence of any reduction of the subluxation with extension of the neck.

The clinical features of the patients are presented in Table 1. Trauma of varying severity was the principal precipitating factor in three patients. The duration of symptoms ranged from 15 days to 5 years (mean 11 months).

All patients were investigated with magnetic resonance imaging, computed tomography, and dynamic radiography. Measurements of structures based on the radiological studies are shown in Table 2. Measurements were made on all available studies and were averaged. Partial or complete occipitalization of the atlas was present in eight patients and fusion of C-2 and C-3 was observed in five. Os odontoideum was seen in three patients.

The surgical techniques employed were described previously. Cervical traction is progressively increased to approximately one-fifth of the total

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body weight prior to induction of anesthesia. The patient is placed prone with the head end of the table elevated to about 35 degrees. Use of the operating microscope facilitates the dissection and screw implantation. The bilateral atlantoaxial facet joints are widely exposed after sectioning of the large C-2 ganglion. Exposure of the facet of the atlas was significantly difficult in the present group of patients because of assimilation of the atlas in most cases and the rostral location of the basilar invagination and the atlantoaxial joint. The joint capsule is excised and the articular cartilage widely removed using the microdrill. The bilateral joints are distracted using the intervertebral spreader used in anterior cervical disc surgery. The status of the dislocation and basilar invagination is evaluated by intraoperative radiography.

The distraction of the C-1 and C-2 facets is maintained by titanium spacers, which are inserted into the joints (Figs. 1 and 2). The size of the spacers depends on the space available within the distracted joint space. The average spacer measures 10 mm in length, 8 mm in breadth, and 4 mm in height. Titanium spacers have multiple small holes to permit bone fusion and are tapered at one end for easier placement in the joint cavity. Multiple bone graft chips are used to fill the distracted joint space around the spacer.

Plate and screw fixation of the region is then carried out by the interarticular technique. A two-holed stainless steel plate measuring 15–20 mm in length and screws of 2.4–2.6 mm diameter and 16–22 mm length are used. The screws are passed bilaterally through the holes in the plate into the lateral mass of the atlas and axis (Figs. 1 and 2). The site and angle of screw implantation is modified to suit the local anatomical situation. The preferred site of screw insertion is at the center of the posterior surface of the lateral mass of the atlas, 1 to 2 mm above the articular surface, but if there was difficulty in exposure due to inadequate space, screw insertion is done through the articular sur-
Fig. 2 Case 1. A: Preoperative radiograph of the craniovertebral region with the neck in flexion showing occipitalization of the atlas, fusion of the C-2 and C-3 vertebrae, atlantoaxial dislocation, and basilar invagination. B: Preoperative radiograph of the craniovertebral region with the neck in extension showing persistent atlantoaxial dislocation. C: Preoperative T1-weighted magnetic resonance (MR) image showing basilar invagination, fixed atlantoaxial dislocation, Chiari I malformation, and syringomyelia. D: Preoperative sagittal computed tomography (CT) scan showing dislocation of the facet of the atlas over the facet of the axis. E: Preoperative CT scan showing basilar invagination, fixed atlantoaxial dislocation, occipitalization of the atlas, and fusion of the C-2 and C-3 vertebrae. F: Postoperative radiograph with the neck in flexion showing the fixation with plates and screws, and the spacers within the joint cavity. G: Postoperative CT scan showing reduction of the basilar invagination and atlantoaxial dislocation. H: Postoperative sagittal CT scan showing reduction of the atlantoaxial dislocation and realignment of the facets. I: Postoperative coronal CT scan showing the screws in the facets of the atlas and axis and distraction of the joint.

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collar for 3 months. All activities involving neck movements are restrained during this period.

Results

All patients improved in the symptoms to varying degrees following surgery and were independent and active in their lives during the follow-up period of 3 to 20 months (mean 7 months) (Table 1). There were no intraoperative or postoperative vascular, neural, or infectious complications. The postoperative changes in various radiological parameters are shown in Table 2. None of the patients suffered delayed neurological deficits requiring transoral or posterior decompression, or any other surgical procedure. No patient needed re-exploration for failure of fixation of the implant. Immediate postoperative and follow-up radiography confirmed fixation and fusion, and reduction of the atlantoaxial dislocation and basilar invagination. Bone fusion was assumed to have occurred if the implant maintained the distraction, and reduction of the atlantoaxial dislocation and basilar invagination on dynamic radiography 3 months after surgery. Successful and sustained reduction of the atlantoaxial dislocation and distraction, and reduction of basilar invagination were observed in all patients. The extent of reduction of the syringomyelia cavity could not be conclusively demonstrated due to the type of metal plate and screws used for fixation. All patients suffered some degree of sensory loss in the distribution of the C-2 nerves.

Discussion

Most patients with syringomyelia related to Chiari malformation but without bony anomaly of the craniovertebral region have hyporeflexia of the upper extremities and spastic lower extremities. Our patients presented with spastic quadriparesis, suggesting that the symptoms were related to compression of the brainstem by the invaginated dens rather than syringomyelia. Patients with fixed atlantoaxial dislocation, basilar invagination, Chiari I malformation, and syringomyelia are relatively younger, with neck pain and far more prominent motor symptoms and ataxia than in patients with similar features without craniovertebral region bony anomaly.

Realignment of the bones in the craniovertebral junction can be attempted in patients with syringomyelia associated with ‘fixed’ atlantoaxial dislocation, with or without basilar invagination and Chiari malformation. We recently described a technique for reduction of fixed atlantoaxial dislocation and distraction, and reduction of basilar invagination. This technique was used to reduce the fixed atlantoaxial dislocation and distraction, and reduce the basilar invagination in patients with associated syringomyelia. We consider that the atlantoaxial joint in such cases is in an abnormal position as a result of congenital abnormality of the bones, and progressive worsening of the dislocation is probably secondary to increasing ‘slippage’ of the atlas over the axis. The slip of atlas over the axis appears to be accentuated by the event of trauma. A history of trauma preceding the clinical events, complaints of pain in the neck, and improvement in neurological symptoms following institution of cervical traction suggest ‘vertical’ instability of the craniovertebral region. Our experience has shown that the joint in such cases is not ‘fixed’ or ‘fused’ but is mobile and in some cases is hypermobile, and is probably the prime cause of the atlantoaxial dislocation and the basilar invagination. The remarkable clinical improvement following reduction of the atlantoaxial dislocation and basilar invagination indicates that the complex of atlantoaxial dislocation, basilar invagination, and syringomyelia is probably secondary to the primary craniovertebral instability.

Opening of the atlantoaxial joint in patients with fixed atlantoaxial dislocation, occipitalized atlas, and basilar invagination is technically relatively complex. The relationship of the vertebral artery to the atlantoaxial joint complex has to be evaluated on the basis of preoperative radiological studies. Wide removal of the atlantoaxial joint capsule and articular cartilage by drilling and subsequent distraction of the joint by manual manipulation provides a unique opportunity to obtain reduction of the basilar invagination and atlantoaxial dislocation. The joints could be maintained in the distracted and reduced position with the help of bone graft and spacers. The subsequent fixation of the joint with interarticular screws and a metal plate provides a biomechanically firm fixation. The fixation was strong enough to sustain the vertical, transverse, and rotatory strains of the most mobile region of the spine. The biomechanical advantage afforded by our technique is illustrated by our successful fusion results in the present and previous series. Following surgery, the alignment of the odontoid process and the clivus, and the entire craniovertebral junction became normalized. We could obtain varying degrees of reduction of the basilar invagination and atlantoaxial dislocation. The atlantoaxial alignments became normal and the tip of the odontoid process receded with respect to Wackenheim’s clival line and Chamberlain’s line suggesting reduction of the basilar invagination and atlantoaxial dislocation.

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al dislocation. The posterior tilt of the odontoid process, as evaluated by the modified omega angle, was reduced after the surgery.\(^{7,14}\)

All patients showed sustained neurological improvement to various degrees, indicating that the procedure was effective. The clinical improvement in the immediate postoperative phase was obviously due to decompression of the invaginated brainstem as shown by postoperative radiology. Stainless steel plates, non-locking screws, and custom-made spacers were used due to the higher costs of branded material. The effect on syringomyelia could not be confirmed as the size of the syrinx could not be radiologically evaluated due to the type of metal instruments used.

Resection of the C-2 ganglion is necessary to achieve the exposure but results in an area of numbness along the distribution of the nerve.\(^{5,9–11}\) However, the area of numbness progressively decreases over the years and was not a disabling feature in any patient. All activities related to neck movements were restricted for a 3-month period to provide an opportunity for bone fusion in the joint.

Experience with the described technique is limited, but our results suggest that realignment and direct fixation of atlantoaxial joint may be indicated for selected patients with syringomyelia. The procedure is technically demanding and requires high anatomic accuracy, but the neurological outcome is extremely gratifying.

References

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Commentary

Drs. Goel and Sharma report their surgical experience in twelve patients with compressive brain stem and high cervical cord lesion secondary to basilar invagination with syringomyelia with mean duration of symptoms being eleven months. Dislocation was manually reducible in all patients without assistance of skeletal traction and/or further extensive disarticulation of the facets and joints for realignment under general anesthesia in a prone position. Their series consists of all selected patients of reducible dislocation. I wonder if they have any experience with patients other than these twelve patients with compressive myelopathy with more advanced irreducible dislocations and longer duration of symptoms. Based on their present experience, how do they manage in such severe advanced cases?

Also, if they could have followed up the syrinx resolution of their patients with serial MR imaging by intraoperative use of instrumentation tools and devices made of titanium instead of steel, they could have provided high core evidence of improvement by resumption of craniovertebral stability by satisfactory alignment. At any rate, the authors are congratulated for their excellent surgical outcome and results in this complex of lesions.

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